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Case report

Sudden death from an asymptomatic phaeochromocytoma: A case report

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ABSTRACT

A case of sudden death due to a phaeochromocytoma in an apparently healthy young man is presented. In cases of sudden death it is necessary to perform a complete and accurate autopsy also looking for alterations in other organs than the heart that could explain the death.

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1. Introduction

Phaeochromocytomas are rare catecholamine-producing tumors that arise from chromaffin cells of the adrenal medulla and extra-adrenal sites (these later called extra-adrenal phaeochromocytomas or paragangliomas); nearly 80–85% of phaeochromocytomas arise from the adrenal medulla, whereas about 15–20% are from extra-adrenal chromaffin tissue.[1,2]

Sporadic forms of phaeochromocytoma are usually diagnosed in individuals aged 40–50 years, while hereditary forms (such as von Hippel-Lindau syndrome, multiple endocrine neoplasia type 1, neurofibromatosis type 1 and familial paragangliomas) are diagnosed earlier; in children it is often extra-adrenal.[3]

Clinical presentation of phaeochromocytoma can vary greatly and it is therefore often referred to as the great mimic; diagnosis is still difficult and often made after death.[4]

2. Case report

2.1. Historical findings

A 43-years old man fell unconscious during a squash training in his own fitness room. Soon admitted to the emergency room, doctors certificated the death. His anamnesis was unremarkable, except for the presence of some episodes of premature ventricular contractions in the last year, no further investigated. An autopsy was performed 24 h after death to understand the cause of death.

2.2. Autopsy findings

External examination showed an apparently healthy man. Massive pulmonary oedema with heavy lungs presenting with pink foam in the main bronchi were detected. Heart (500 g in weight) showed mild hypertrophy. Coronary arteries presented moderate lipoidosis. Abdominal organs were normal; the left adrenal gland was moderately enlarged compared with the right one, without a distinctly appreciable mass.

2.3. Histological findings

Myocardium evaluation presented a moderate myocytic hypertrophy and a lot of small areas of fibrosis with poor cells (Fig. 1). In the medulla of the left adrenal gland, cells with round nuclei and prominent nucleoli, arranged in nests bound by a delicate fibrovascular stroma, were found (Fig. 2). Immunohistochemically, these cells strongly and diffusely stained with antibodies against synaptophysine (Fig. 3A–B).

2.4. Toxicological findings

Urine and blood samples were taken on autopsy. Ethanol on blood and illicit drugs (cocaine, opiates, amphetamines, cannabinoids) on blood and urine were negative.

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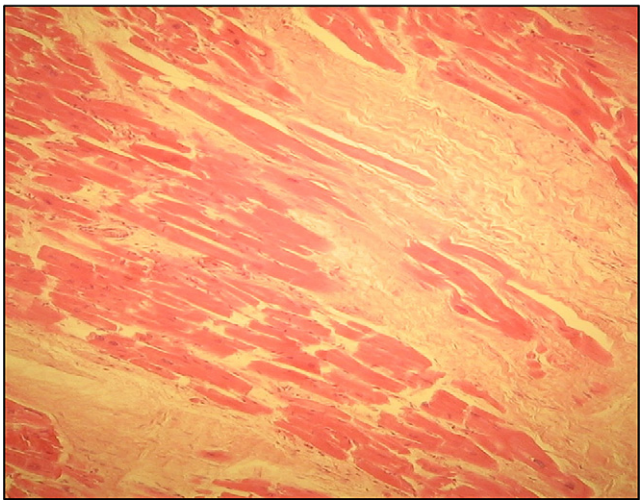

Fig. 1. Myocardium: moderate myocytic hypertrophy and spot of fibrosis with poor cells (HE stain, 100X).

3. Discussion

Diagnosis of phaeochromocytoma is very difficult since this tumor gives poor signs of its presence; sometimes its clinical presentation can be consequence of a blunt abdominal trauma.[5,6] Although difficult, omission in the diagnosis can have medico-legal implications.[7]

In this case there were apparently no clinical signs of the tumors, except for the presence of some premature ventricular contractions; the man was in apparently good health and dedicated regularly to sport activity. Cardiac arrest during a squash training suggests that sympathetic iperactivity played a role in the hyper-secretion of hormones from the tumor with consequent activity on the heart. The presence of pulmonary oedema gives rise to the hypothesis of an acute cardiac failure, maybe due to a fatal arrhythmia.

In case of sudden death, the pathologist's attention is toward the heart usually. This case underlines that it is necessary to consider causes of death other than the heart.[8,9]

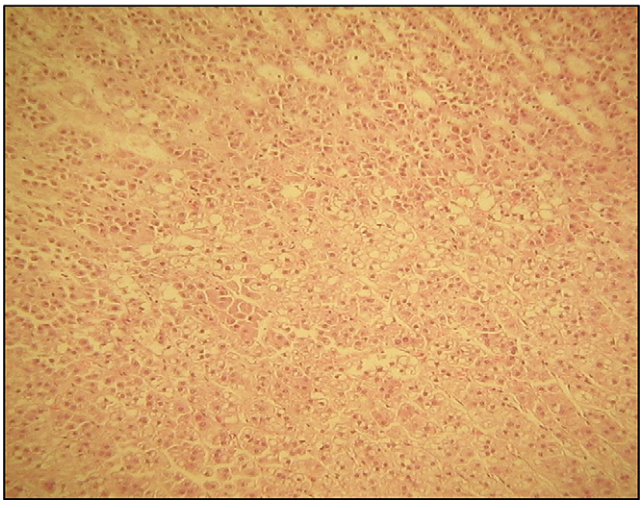

Fig. 2. Left adrenal gland: cells with round nuclei and prominent nucleoli, arranged in nests bound by a delicate fibrovascular stroma (HE stain, 25X).

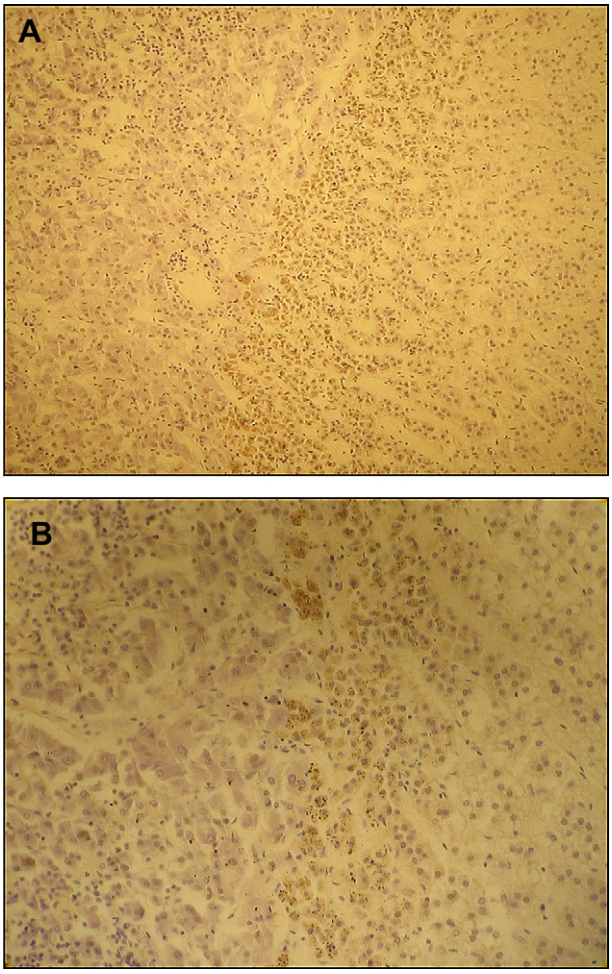

Fig. 3. (A–B). Left adrenal gland: cells strongly and diffusely stained with antibodies against synaptophysine (A-25X; B-100X).

Conflict of interest

None.

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